A Rare Bednar Tumour Of Neck

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Abstract

Bednar tumour (pigmented Dermatofibrosarcoma protuberans) is a variant of dermatofibrosarcoma protuberans(DFSP)-constitute 5% of all DFSP-a relatively uncommon soft tissue neoplasm with intermediate to low grade malignancy rarely metastasizing to regional lymph nodes or distant sites but prone for local recurrence.

We report here in a case of 68year old female who presented with a huge recurrent Bednar tumour(40cm X 36cm x 26 cm) arising from neck region after a gap of five years of primary excision.

Review of previous studies and histopathological examination of recurrent lesion was suggestive of Bednar tumour. We performed a wide excision with 3 cm free skin margins. Soft tissue reconstruction was done to close the defect at the resected area.Recognition of this tumour is important because of excellent prognosis after adequate surgical excision.

INTRODUCTION

Pigmented dermatofibrosarcoma protuberans or previously known as Pigmented storiform neurofibroma is a rare variant of DFSP ,described by BEDNAR in 19571.The histological picture shows melanin laden dendritic cells with spindle shaped cells in storiform pattern.The tumour mostly occurs on trunk,upper and lower extremities,few in head and neck region2..It is a slow growing,locally invasive-painless cutaneous multilobulated lesion, burn or surgical scar3 at the initial stage often the cause.

It has been shown that Giant cell Fibroblastoma has a close histogenic relationship to Dermatofibrosarcoma protuberans4 and it has been reported that Giant cell fibroblastoma can transform into DFSP5-7.We present here a case of massive Bednar tumour of neck region presenting five years after initial excision.

CASE REPORT

A 68 year old female was admitted with a huge painless nodular tumour

over the neck region. She gives history of insect bite at the same spot eight years

back following which a tumour developed over next one year slowly growing upto

four cm in diameter. The lesion was excised under local anaesthesia and histological

diagnosis revealed Dermatofibroma. She had another recurrence of the tumour two

year following the first excision which she neglected and the tumour gradually

progressed to a very large size.

The patient denied fever , chills, bony pain or any other constitutional

symptoms. On physical examination ,patient was not cachectic with no palpable

lymphadenopathy.Respiratory, CVS and GI exam was within normal limits.

Skin examination revealed 40 x 34 cm exophytic mass over neck region with

multiple soft protuberances of varying size none exceeding 8 cm,soft in consistency

with thin skin cover over certain areas with ulceration. The nodules appeared fixed

to the overlying skin but were mobile over the deep tissue.There was no other

positive finding on complete head and neck examination and there was no other

swelling elsewhere in the body.

Lab. Findings were within the normal limits.Chest X-ray was WNL.

Computed tomography (CT) revealed no invasion of the mass into deeper

structures .

A wide and deep excision with 3 cm margins of normal tissue all around

the lesion was performed. Grossly,the resected specimen measured 40 X 36X26 cm diameter with multiple discrete nodular mass,the greatest being 8 cm in diameter. The nodules were well circumscribed,had a firm consistency and showed a whorled pattern on cut section.

On histologic examination ,they were found to be composed of a uniform

population of fibroblast like cels,arranged in a storiform pattern . The cells had hyperchromatic oval to spindle shaped nuclei with low mitotic activity.The striking feature was presence of bipolar and multipolar dendritic cells with tentacle like processes emanating from nucleus .

Immunohistochemical study showed positive reaction to CD 34 and

VIMENTIN and negative for protein S-100 but melanin containing cells showed

positive for protein S-100.

The resected specimen showed tumour free margins. Soft tissue

reconstruction was done at the resected site of tumour.

Figure 1

Macroscopic view of tumour located on anterior part of chest wall.

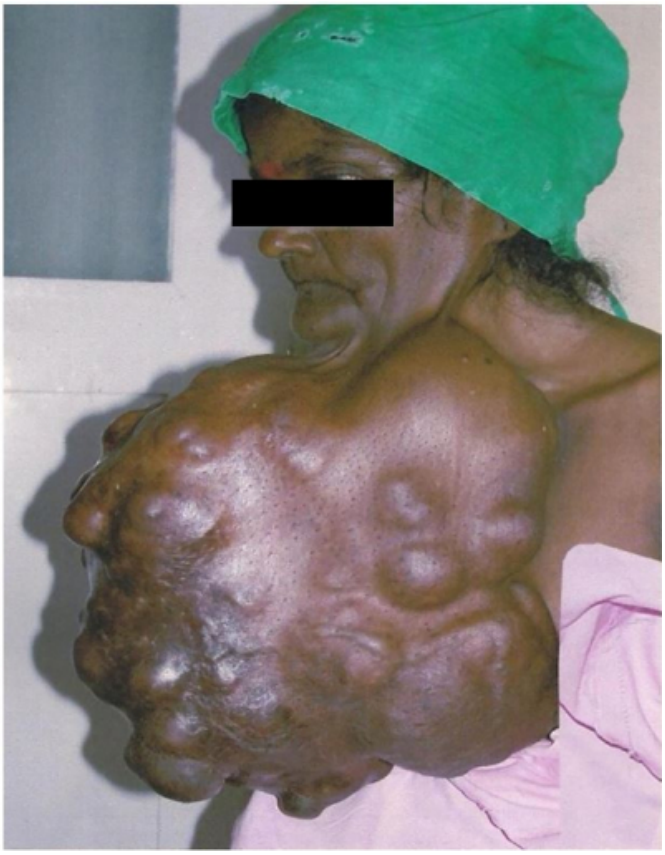


Figure 2

Postoperative view of the resected Specimen (40 x 36 x 26 cm)

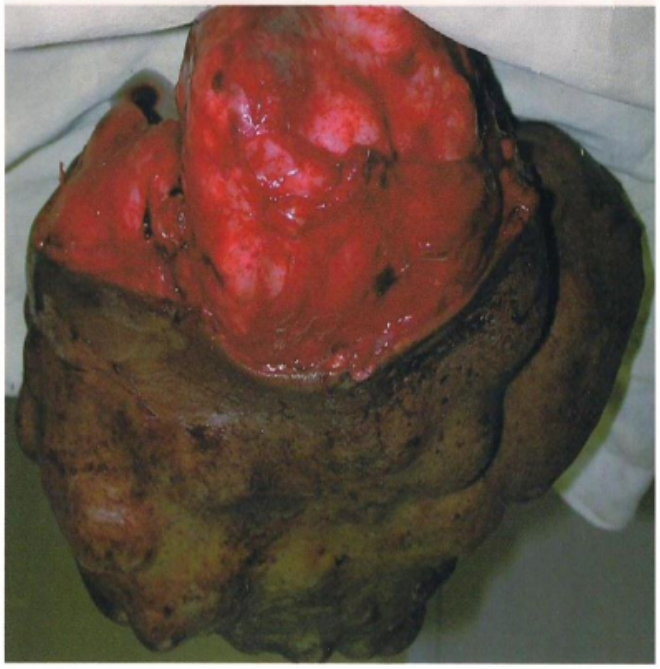


Figure 3

Neoplasia occupying the dermis and compose of fusiform cells and cells containing melanin

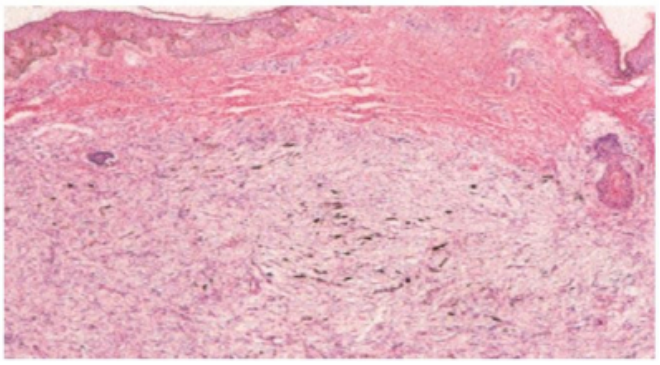


Figure 4

Melanin pigment seen in cells of dermis and Subcutaneous layer

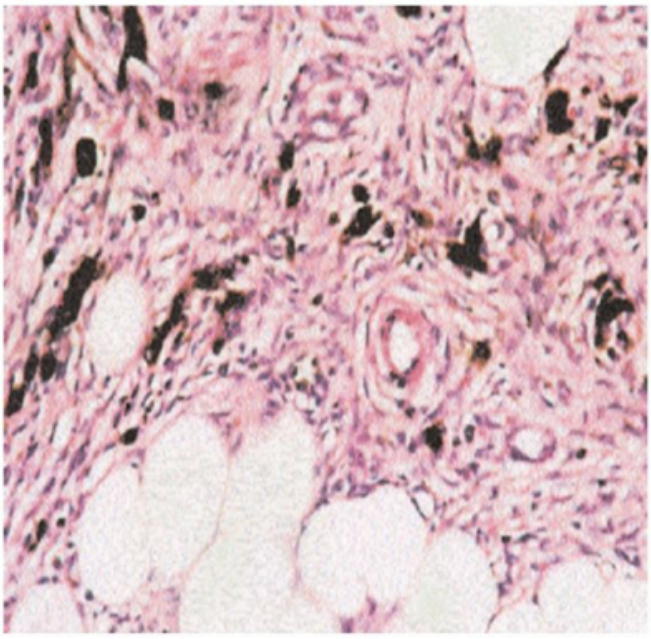
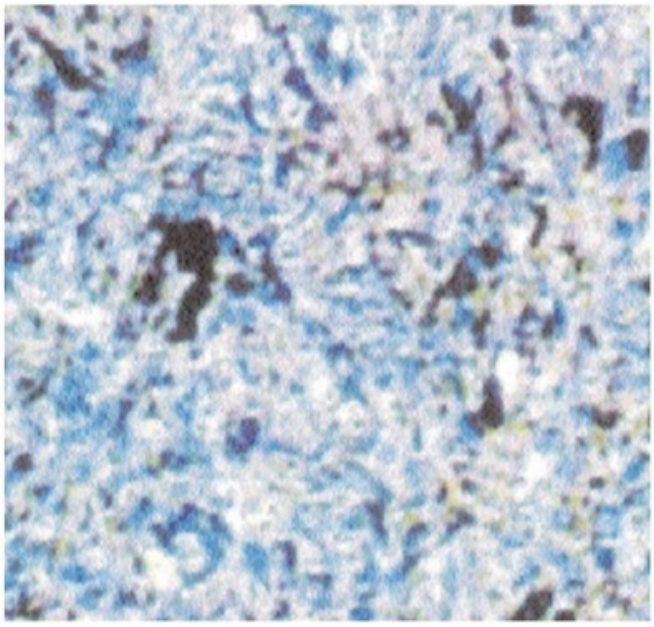


Figure 5

Immunohistochemistry—Positivity for antibody against CD 34,which confirmed the diagnosis



Retrograde histological re-examination of the specimen obtained from the second excision of the recurrent tumour showed high expression of CD34 and revealed present tumour was actually a recurrence of this tumour.

The wound healed without any complication and the post operative course was uneventful,with no paraesthesia or skin sloughing at the reconstruction site.

DISCUSSION

The case presented here was DFSP ,which was initially mistaken to be

dermatofibroma.Dermatofibromas composed predominantly of fibroblasts extending

into the subcutaneous tissue are difficult to distinguish from dermatofibrosarcoma

protuberans8 though the patterns are different.

Bednar tumour account for less than 5 % of all DFSP9 and more common

in Black male population .Different theories have been given regarding origin of

Pigment laden cells .Dual cell origin suggests CD 34

positive spindle cells of

Mesenchymal and pigmented cells of neuroectodermal origin10.

In the case reported here correct clinical diagnosis was not reached initially

but only later on recurrence after subjecting to histopathological and

immunohistochemical studies.Condensation of connective tissue at the periphery

may give a false appearance of encapsulation but tumour may extend well beyond

margins11. We need to be aware of this condition12 and confirm histological

diagnosis before excision.

Recurrence are due to inadequate excision with tumour extending to deep

resection margin. With recurrence the lesion becomes less well differentiated13 and

chances of metastasis increases.Though rare,dissemination occurs by hematogenous

route and rarely lymphatically . The principal site being lungs though

bones,liver,pancreas,stomach,intestine,thyroid and brain may be involved

Surgery with a safety margin of 3 cm including the underlying fascia

serves good with Computerized tomogram helpful in deciding the line of incision to

prevent recurrence or metastasis.Mohs micrographic surgery14 has maximum

oncologic effectiveness and is the accepted treatment of choice.

Chemotherapy is not used in treatment while radiotherapy can be used as

adjunct to surgery in cases with positive resection margins.Molecular targeted

therapy and Imatinib may provide alternative treatment to unresectable tumours or

adjunctive treatment in addition to surgery.

In short ,Bednar tumour is a rare tumour.Painless ,cutaneous and

multilobulated lesion should arouse suspicion of this tumour and core or incision

biopsy should aim at pre-operative histologic diagnosis.Adequate excision will avoid

recurrence and excellent prognosis.

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